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Teaching cases

Follicular dendritic cell sarcoma of the tonsil

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ABSTRACT

Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm that occurs extranodally and nodally. The following case report describes a 24-year-old male patient who suffered from FDCS of the tonsil. He presented at the ENT Department of the University Hospital Magdeburg with throat pain that had lasted for 3 months. There were neither B symptoms nor abnormal fatigue. An extended tonsillectomy was performed. The morphological and immunohistochemical findings confirmed the diagnosis of FDCS.

FDCS should be considered as an important differential diagnosis in spindle cell tumors of the tonsil.

Introduction

Clinical history

A 24-year-old male patient was admitted to the University Clinic of Magdeburg with a complaint of throat pain which, according to him, had lasted for almost 3 months. B-symptoms or abnormal fatigue were not reported by the patient. Before admission to the hospital, he had visited an otolaryngologist who noticed an asymmetry of the tonsils, and performed a biopsy of the left tonsil. The histopathological examination of the biopsy revealed a follicular dendritic cell sarcoma. At the time of the admission to the hospital, intraoral examination showed a massive swelling of the left tonsil (Fig. 1). This tonsil was found to be smooth, and exhibited easy luxation. No necrotic or ulcerated areas were observed. The tonsil of the opposite side showed signs of chronic tonsillitis and appeared scarred. In addition, the left tympanic membrane showed a retraction pocket and was atrophic. Neck examination revealed two lymph nodes of approximately 1 cm in size on both sides of the neck in region II. The results of a general ear nose and throat examination were normal without any relevant comorbidity.

Diagnostics

Ultrasonography revealed a lymphadenitis that was more severe on the left side of the neck than on the right side. A tumor mass of approximately 24 mm in diameter with a homogenous structure and low echogenocity (Fig. 2) was found in the left tonsillar region.

The MRI scan showed a mass of 25 mm × 20 mm in the area of the left tonsil, which limits the space of the left oropharynx (Fig. 3).

The native 18-fluoro-desoxy-D-glucose-positron-emission-tomography-computed-tomography (F-18-FDG-PET CT) scan showed only a slight increase of the glucose metabolism in the area of the left tonsil. No sign of metastasis was found in other parts of the body; however, their presence could not be ruled out with absolute certainty.

Therapy

Extended laser-tonsillectomy was performed with a CO_2-laser on the left tonsil, where the tumor was located, under general anesthesia. The margins were free of tumor. Furthermore, conventional tonsillectomy was also performed on the opposite side, as requested by the patient.

Moreover, the patient was treated with cefuroxime for 10 days after the operation. Adjuvant radiochemistry or neck dissection was not performed. Nevertheless, the patient was requested to return to the clinic for further regular examinations. Until now, no residual tumor mass has been detected.

General information on the disease

Follicular dendritic cells (FDCs) are non-phagocytic, non-lymphoid antigen-presenting cells of the accessory immune system. These cells of mesenchymal origin are capable of

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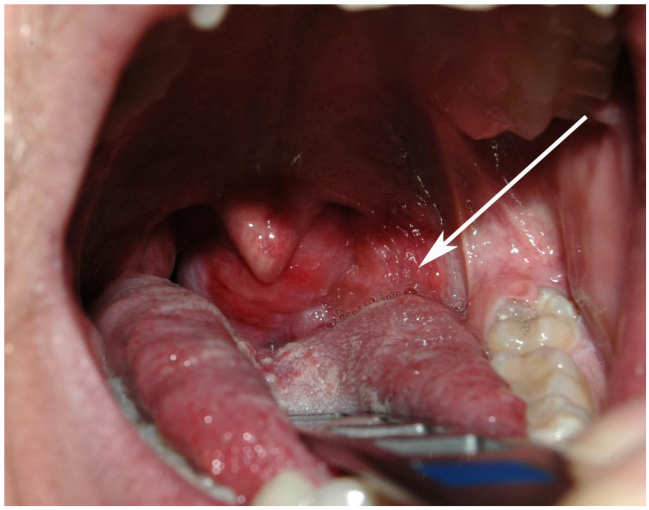

Fig. 1. Intraoral examination with the tumor of the left tonsil.

capturing, processing and presenting antigens and immune complexes to T-cells and B-cells in order to maintain the humoral immune response. They are part of the dendritic cell group, which can be divided into four main categories found in lymphatic tissue: interdigitating dendritic, indeterminate, Langerhans and follicular dendritic cells [6]. FDCs are mainly found in the germinal centers of primary and secondary follicles of lymph nodes, the spleen or mucosa associated lymphatic tissue (MALT) within the network of other local cells.

The first case of a FDC sarcoma was described by Lennert [8]. Then, in 1986, Monda et al. reported the first case of primary neoplasm of FDCs in the lymph node [9]. To date, only a few cases of FDC sarcoma in the tonsil have been described [13]. Few extra nodal FDCS were reported by Youens and Waugh [14], and in the pharyngeal or abdominal region [12].

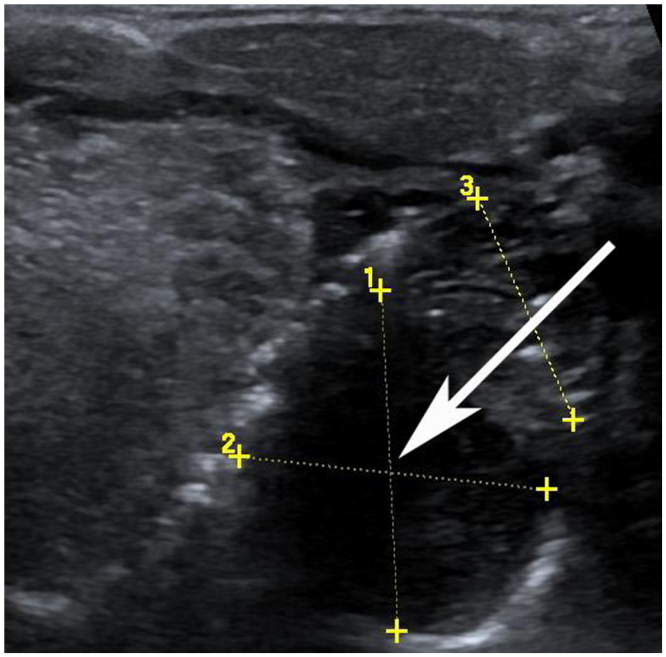

Fig. 2. Ultrasonography of the left tonsil depicting the tumor mass of 24 mm diameter.

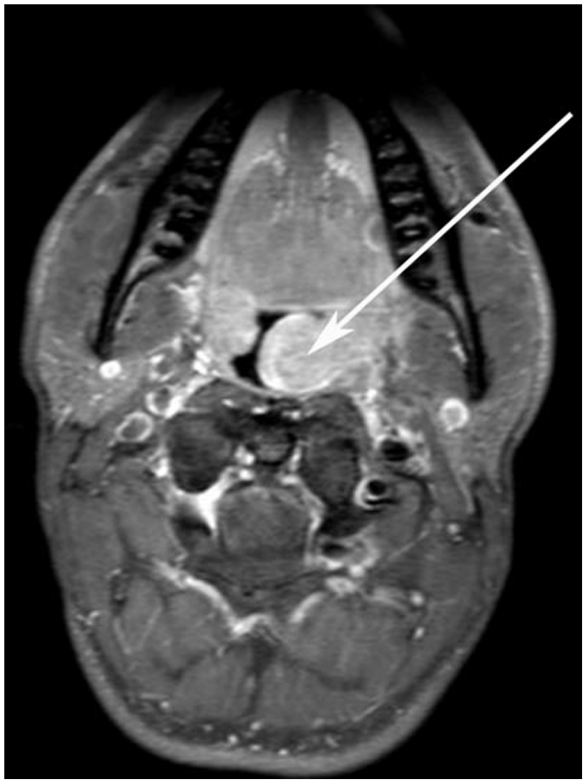

Fig. 3. MRI scan of the head with the tumor of the left tonsil.

Material and methods

Histology

After complete tonsillectomy, the tissue of the left tonsil was collected and preserved in buffered formalin (10%). Subsequently, the tissue was dehydrated and embedded in paraffin. Sections (3.0 μm thick) from formalin-fixed, paraffin-embedded tissue were stained with hematoxylin and eosin (HE). Light microscopy was performed using an Axioplan microscope (Zeiss, Oberkochen, Germany) and a Hamamatsu Nanozoomer (Hamamatsu Photonics, Hamamatsu, Japan).

Immunohistochemistry

Sections (3.0 μm thick) from formalin-fixed, paraffin-embedded tissue were incubated with a panel of standard antibodies against cytokeratins (MNF116, AE1/AE3), epithelial membrane antigen (EMA), vimentin, sm-actin, desmin, S-100 protein, HMB45, leukocyte common antigen (LCA), KiM4p, CD23, and ki-67 using a Benchmark XT (Ventana Medical Systems, Tucson, USA). The reactions were visualized using DAB detection.

Results

Morphology and immunophenotype

The tumor presented as a well-defined mass in an otherwise regularly structured tonsil (Fig. 4). The spindled tumor cells formed fascicles and storiform patterns. The nuclei showed dispersed chromatin and small nucleoli without significant cytological atypia. Only a few mitoses were observed (4/10 HPF). Small lymphocytes were scattered across the tumor. The surgical margins were tumor-free.